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Radiology Corner Case #16

Dextroposition of the Heart

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History

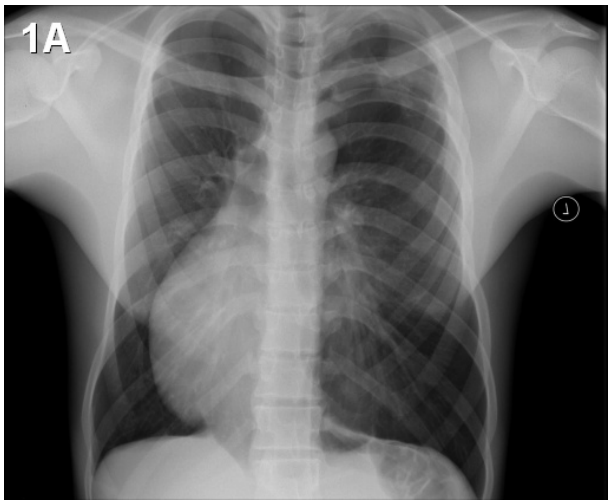
An 18-year-old male recruit underwent a physical examination to assess fitness for enrollment into the Naval Academy. There was no health complaint. His medical history was significant for a pectus excavatum. His surgical history was notable only for a cosmetic repair of the pectus at age 14. Review of systems included an ability to participate in athletics without limitation. His admission physical exam documented only "a left chest wall 6 cm scar consistent with surgical history." The screening chest x-ray is presented below (Fig 1A). Technical limitations impeded a thorough echocardiographic evaluation and a cardiac magnetic resonance imaging (MRI) study was performed to determine whether the condition represented dextroposition, true mirror-image dextrocardia, or a more sinister culprit such as corrected transposition of the great arteries (Fig. 1B, coronal; Figs. 1C, 1D and 1E, axial cranial to caudal).

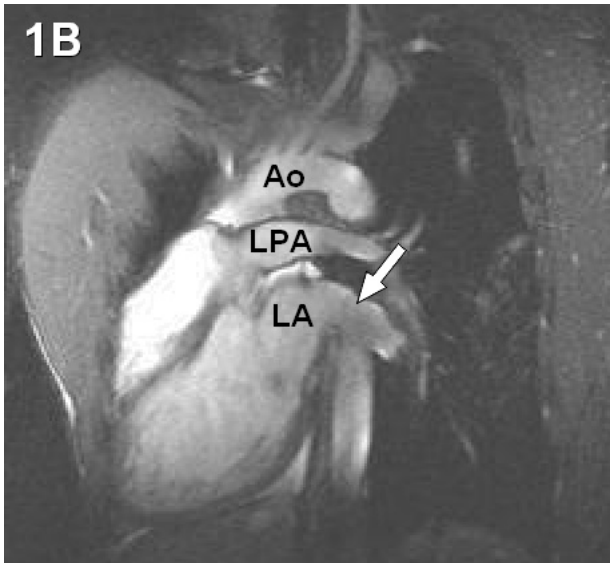
Imaging Findings

The PA radiograph (Fig. 1A) demonstrates a right-sided heart. Cardiac MRI images further define the internal cardiac anatomy. On a coronal bright blood MRI image (Fig. 1B; LA = left atrium; LPA = left pulmonary artery; Ao = aorta), the pulmonary veins (arrow) are seen entering into the left-sided atrial chamber (i.e. left atrium). This confirms the usual atrial position. On axial black blood MRI images (Figs. 1C thru 1E, cranial to caudal), the moderator band (arrow, Figs. 1D and 1E; RV = right ventricle; LV = left ventricle), a structure unique to the right ventricle, is visualized in the right-most ventricle in Fig. 1C, confirming a normal d-loop ventricular development and orientation. On a more cranial axial cardiac MRI image (Fig. 1C), the aorta (Ao) is noted to be posterior and to the right of the pulmonary artery (PA). Again, normal anatomy is confirmed. These findings are consistent with normal intra-cardiac orientation. Since internal cardiac anatomy is normal in spite of the organ's right-sided location, the diagnosis is dextroposition of the heart.

Diagnosis

Dextroposition of the Heart





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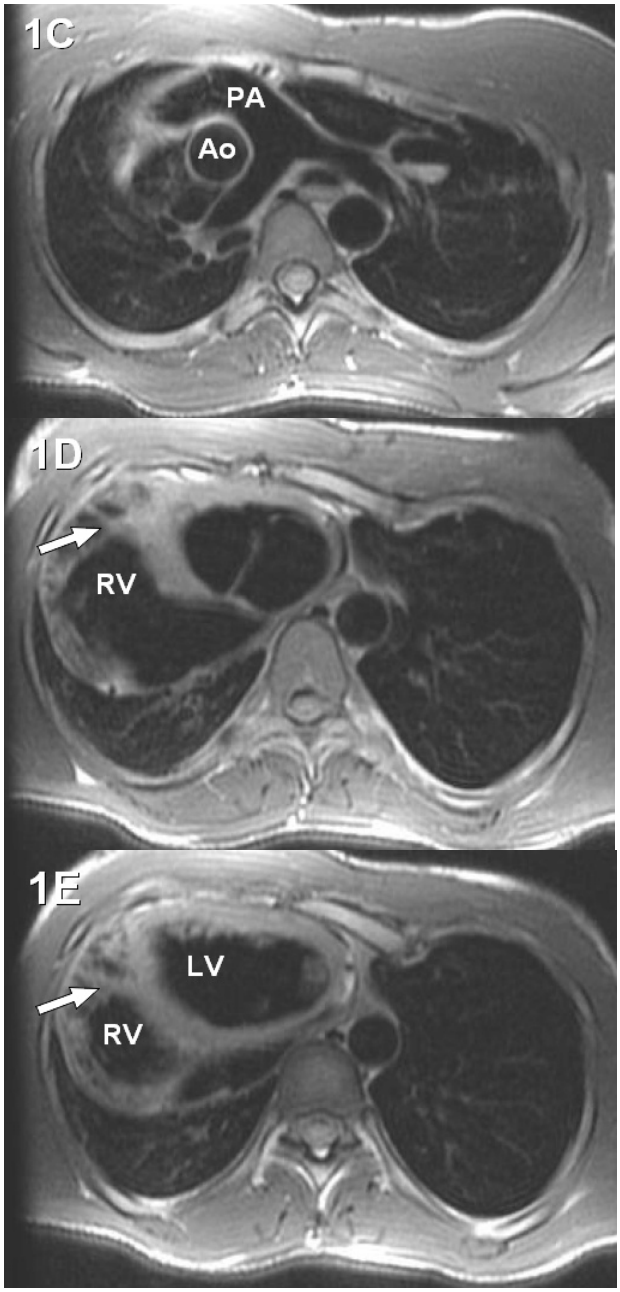


Discussion

On the basis of 1960s-era studies of necropsy specimens, Van Praagh et al.[1] developed a system by which to classify conditions causing right-sided heart. Entitled "segmental analysis," this system is useful to the present in the diagnosis of congenital cardiac disorders. The determination of the precise location and relationships of the cardiac chambers and great vessels is the foundation of this endeavor.[2-8] While Van Praagh et al. relied on gross anatomical patterns in their studies, the combined modalities of electrocardiography (EKG), echocardiography, and x-ray have allowed the application of segmental analysis to living subjects. Performance of echocardiography on structurally and positionally abnormal hearts is often difficult and subject to misinterpretation. On the other hand, cardiac MRI can provide the segmental data with relative ease.

Segmental Analysis

A. Location of the Atria

Determination of the orientation of the atria is the first step in segmental analysis. A universal rule exists which makes atrial location easy. Because of a fixed developmental pattern in early embryogenesis, atrial situs is almost always concordant with visceral situs. In other words, the left atrium is always found on the same side as the stomach. The right atrium stays in parallel with the liver. Locating the abdominal organs by physical exam or x-ray thus provides atrial situs. The term "solitus" means "usual." Thus "situs solitus" refers to normal position of the atria. "Situs inversus" describes mirror-image reversal of the atria and the visceral contents (Fig. 2).

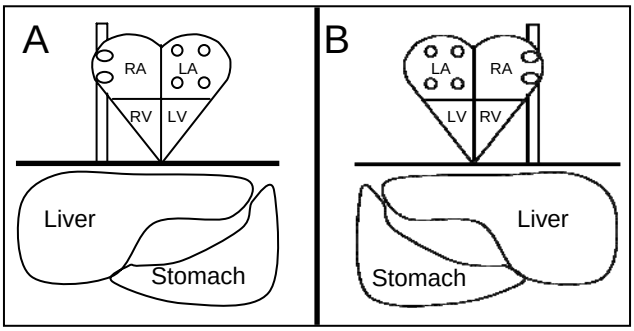


Figure 2. Atrial-visceral relationships. A) Situs solitus, B) Situs inversus

Constant anatomical relationships exist within the heart as well. The atrial chamber that is connected to the inferior vena cava is typically the right atrium. The pulmonary veins typically empty into the left atrium. Identifying these structures by echocardiography or cardiac MRI yields atrial situs. In this case, abdominal exam, chest x-ray (Fig. 1A) and cardiac MRI (Fig. 1B) confirm situs solitus.

B. Location of the Ventricles

The primitive heart tube of early embryology develops a rightward turn in the first month of development. The right-sided bulbous cordis becomes the right ventricle and the posterior, left-sided "ventricle" becomes the left ventricle. This normal anatomical relationship is termed dextro-loop (d-loop). If the primitive heart tube instead develops leftward looping (l-loop), the right ventricle develops on the left and vice versa. Mirror image symmetry is manifested.

The determination of ventricular location is best achieved by direct visualization. Current literature recommends echocardiography localization of the tricuspid and mitral valves. The valves exist, as a general rule, in concordance with their associated ventricles. In our patient, the "technically-limited" echocardiography included no comment on ventricular loop.

Cardiac MRI, as shown in this case, often provides the answer. The presence of a moderator band and infundibulum identifies the morphologic right ventricle and enables the

identification of the right ventricle.[9] Once identified, the right ventricle location and orientation confirmed the normal d-loop development of the ventricles in the current case (Figs. 1D and 1E).

C. Location of the Great Arteries

The great arteries are either normally related to the ventricles or they are transposed. Great artery relationships can be simplified by consistently visualizing them in axial views at their ventricular origin (Fig. 3).

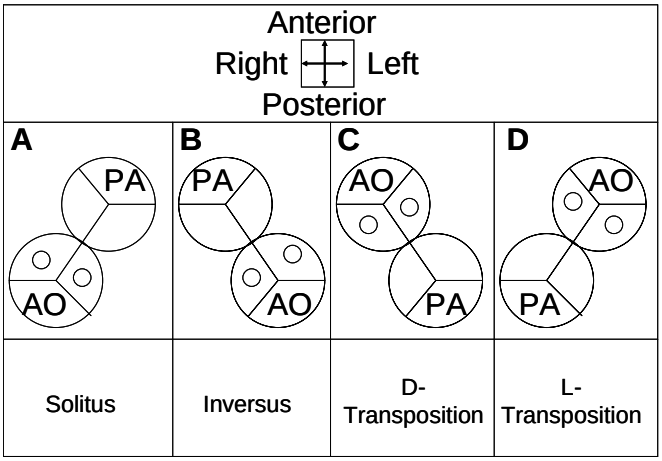
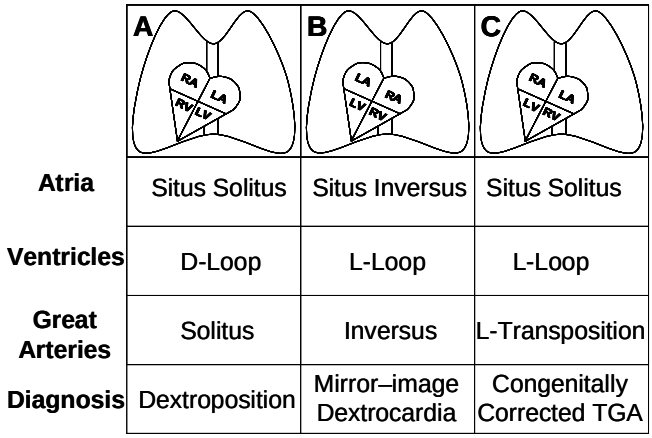


Figure 3. The four potential relationships between the great arteries (i.e. aorta [AO] and pulmonary artery [PA]).
A) Solitus relationship (usual or normal relationship) is present when the aortic valve is posterior and to the right of the pulmonary artery.
B) Inversus relationship is where the aortic valve is posterior and to the left of the pulmonary valve (mirror-image of the usual or normal arrangement).
C) Complete transposition (d-transposition) is present when the aortic valve is anterior and to the right of the pulmonary valve.
D) Congenitally-corrected transposition (l-transposition) is present when the aortic valve is anterior and to the left of the pulmonary valve.

A posterior and rightward aorta in relation to the pulmonary artery is the norm. When an l-loop develops, it "pushes" the anatomical left ventricle right-ward. It is appropriate for the aorta to maintain its "opposite" anatomical relationship. Just as the ventricles assume a mirror-image of normal, so too do the great vessels. The aorta remains posterior but is positioned leftward of the pulmonary artery. The configuration, seen in mirror-image dextrocardia, is called "inversus." In transposition, the aorta originates from the right ventricle. This is visualized as an anterior aorta in comparison to the pulmonary artery. Likewise, the left ventricle is connected to the pulmonary artery. Transposition of the arteries in a normally-configured d-loop ventricular arrangement is termed d-transposition. The equivalent in patients with l-loop ventricles is l-transposition.

Practically, localization of the great arteries is done by direct visualization. Prior to the development of current advanced cardiac MRI techniques, echocardiography was used primarily. Angiography is an alternative option. However, as shown in this case, MRI clearly demonstrates solitus relationship of the great arteries (Fig. 1C).



	A	B	C
Atria	Situs Solitus	Situs Inversus	Situs Solitus
Ventricles	D-Loop	L-Loop	L-Loop
Great Arteries	Solitus	Inversus	L-Transposition
Diagnosis	Dextroposition	Mirror–image Dextrocardia	Congenitally Corrected TGA

Figure 4. Common causes of right-sided heart

Summary and Significance

The Cardiac MRI findings allow the following conclusions:

1. The heart is in the right hemi-thorax.
2. Atrial situs is solitus.
3. The ventricles are positioned according to d-loop development.
4. The great arteries demonstrate solitus development.

These findings are consistent with normal intra-cardiac orientation. In other words, the patient has dextroposition. His normal heart is shifted into the right hemi-thorax. Dextroposition is associated with hypoplastic right lung and deformities of the chest wall and diaphragm. Indeed, the patient's pectus excavatum indicates a developmental problem late in embryology.[1-3] This case illustrates the ease with which cardiac MR can elucidate complicated anatomy. Few references are available on this topic [2-9], but the application of cardiac MR to segmental analysis is intuitive.

The most common causes for right-sided heart are shown in Figure 4. The associated segmental analysis is provided for each. In this case, the MRI examination ruled out congenitally-corrected transposition of the great arteries. This condition can exist, undiagnosed, into adulthood. Diagnosis is critical for future management.

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